B4

26. (Amended) The method of Claim 14 wherein said treatment with human recombinant α-L-iduronidase reduces lysosomal storage caused all or in part by said deficiency in α-L-iduronidase.

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28. (Amended) The method of Claim 14 wherein said treatment results in increase in percent forced vital capacity, increase in distance of six-minute walk, reduction of liver volume and urinary glycosaminoglycan excretion, reduction in spleen size and apnea/hypopnea events, increase in height and growth velocity in prepubertal patients, increase in shoulder flexion and elbow and knee extension, reduction in symptoms related to cardiac function, and increase in endurance and reduction of limitations of daily activities.

The following new claims are added:

29. (New) A method of treating diseases caused all or in part by a deficiency in α-L-iduronidase, comprising the steps of:

(a) administering a pharmaceutical composition comprising a purified human recombinant α-L-iduronidase, or biologically active fragment or mutein thereof, having a purity of greater than about 99%, to a human subject in need thereof; and

(b) optimizing said treatment by evaluating biochemical and clinical symptoms of said subject through routine assessment of history, physical examination, echocardiography, electrocardiography, magnetic resonance imaging, polysomnography, skeletal survey, range of motion measurements, corneal photographs, and skin biopsy.

B6

30. (New) The method of Claim 29 wherein the disease is mucopolysaccharidosis.

31. (New) The method of Claim 29 wherein the disease is mucopolysaccharidosis I.

32. (New) The method of Claim 29 wherein the disease is selected from the group consisting of: Hurler's disease, Scheie syndrome and Hurler-Scheie syndrome.

33. (New) The method of Claim 29 wherein said subject suffering from the disease demonstrates about 1% or less of a normal α-L-iduronidase activity.

34. (New) The method of Claim 29 wherein a dose of at least about 125,000 units or 0.5 mg/kg of said human recombinant α-L-iduronidase is administered weekly to a patient suffering from a deficiency thereof

35. (New) The method of Claim 29 wherein said administering is a slow infusion of at least 0.5 mg/kg of said formulation for about an hour, followed by a rapid two-hour infusion rate.

36. (New) The method of Claim 29 wherein said infusion is used to minimize complement mediation clinical allergic reactions.

37. (New) The method of Claim 29 wherein said treatment with human recombinant α-L-iduronidase reduces lysosomal storage caused all or in part by said deficiency in α-L-iduronidase of said human subjects .

38. (New) The method of Claim 29 wherein said treatment results in normalization of liver volume and urinary glycosaminoglycan excretion, reduction in spleen size and apnea/hypopnea events, increase in height and growth velocity in prepubertal patients, increase in shoulder flexion and elbow and knee extension, and reduction in tricuspid regurgitation or pulmonic regurgitation.

39. (New) A method of treating diseases caused all or in part by a deficiency in α-L-iduronidase, comprising the steps of:

administering a pharmaceutical composition to a human subject in need thereof; wherein said pharmaceutical composition comprises a purified human recombinant α-L-iduronidase, or biologically active fragment or mutein thererof, having a purity of greater than about 99%.

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40. (New) The method of Claim 39 wherein the disease is mucopolysaccharidosis.

41. (New) The method of Claim 39 wherein the disease is mucopolysaccharidosis I.

42. (New) The method of Claim 39 wherein the disease is selected from the group consisting of: Hurler's disease, Scheie syndrome and Hurler-Scheie syndrome.

43. (New) The method of Claim 39 wherein said subject suffering from the disease demonstrates about 1% or less of a normal α-L-iduronidase activity.

44. (New) The method of Claim 39 wherein a dose of at least about 125,000 units or 0.5 mg/kg of said human recombinant α-L-iduronidase is administered weekly to a patient suffering from a deficiency thereof.

45. (New) The method of Claim 39 wherein said administering is a slow infusion of

at least 0.5 mg/kg of said formulation for about an hour, followed by a rapid two-hour infusion rate.

46. (New) The method of Claim 45 wherein said infusion is used to minimize complement mediation clinical allergic reactions.

47. (New) The method of Claim 39 wherein said administering with human recombinant α-L-iduronidase reduces lysosomal storage.

48. (New) The method of Claim 39 wherein said administering results in a decrease in the volume of the liver of said patient by at least 5%.

49. (New) The method of Claim 48 wherein said administering results in a decrease in the volume of the liver of said patient by at least 19%.

50. (New) The method of Claim 39 wherein said administering results in a decrease in the volume of the spleen of said patient by at least 13%.

B6 cont

51. (New) The method of Claim 39 wherein said administering results in a decrease in the urinary glycosaminoglycan excretion of said patient by at least 60%.

52. (New) The method of Claim 39 wherein said patient is a prepubertal patient and said administering results in an increase of the height growth velocity of said patient by at least 2.4 cm/year.

53. (New) The method of Claim 39 wherein said patient is a prepubertal patient and said administering results in an increase of the weight growth velocity of said patient by at least 2.4 kg/year.

54. (New) The method of Claim 39 wherein said administering results in an increase of the shoulder flexion of said patient.

55. (New) The method of Claim 39 wherein said administering results in an increase of the elbow and knee extension of said patient.

56. (New) The method of Claim 39 wherein said administering results in a reduction of apnea and hypopea events of said patient.

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57. (New) The method of Claim 39 wherein said patient has tricuspid regurgitation or pulmonic regurgitation caused all or in part by a deficiency in α-L-iduronidase treatment and wherein said administering results in a reduction in said tricuspid regurgitation or pulmonic regurgitation.

REMARKS

The Amendments

Claim 19 is amended to recite "mucopolysaccharidosis". Amendment is made to further clarify that the abbreviation "MPS" refers to "mucopolysaccharidosis". Support for the amendent is found, for example, in page 5, line 25.

Claim 21 is amended to recite "said human subject". Amendment is made to further clarify that "said subject" refers to "said human subject" found in Claim 14. Support for the amendent is found, for example, in Claim 14 as originally filed.

Claim 22 is amended to recite "said human subject suffering from said deficiency". Amendment is made to further clarify that "patient" refers to "said human subject", and "a deficiency" refers to "said deficiency", found in Claim 14. Support for the amendent is found, for example, in Claim 14 as originally filed.

Claim 24 is amended to recite "a slow infusion". Amendment is made to further clarify that the phrase "slow infusion" does not have antecedent support. Support for the amendent is found, for example, in Claim 24 as originally filed.